normal in both. Both of them had extracardiac malformation: one had oesophageal atresia (requiring surgery at day 1), the second had malformation of the ear and micropenis. Both had neonatal PAH. The first patient received bosentan at the second month of life. He had persistent PAH (PA pressure of 119/48, mean 77 mm Hg) and underwent closure of a small VSD at the age of 7 months. Two years and half after surgery, he remains in NYHA class II with similar PA pressure despite bosentan therapy. He is awaiting for combination therapy with sildenafil. The second presented pulmonary distress at birth requiring mechanical ventilation. He was placed immediately under a combination therapy of epoprostenol and NO for 4 days, received then treprostinil for 15 days and sildenafil. He could be weaned off the ventilator after 3 days. One month and half after birth, he is doing well under sildenafil but has persistent moderated PAH (tricuspid regurgitation velocity of 3 m/s).

The antenatal existence of high grade arteriovenous shunt could explain high pulmonary artery pressure. However, the persistence of PAH in these patients remains unclear. In combination with research of chromosomal anomaly and congenital malformation, infants with agenesis of the ductus venosus should benefit after birth of serial ultrasound examination to rule out PAH.

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Conotruncal and coronary artery development in two mouse models of congenital heart defects

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Conotruncal heart defects are among the most frequent congenital heart diseases. Coronary artery anomalies are commonly associated with outflow tract malformations. The molecular and cellular mechanisms underlying their development have yet to be unravelled. TBX1, encoding a T-box transcription factor, is the major DiGeorge syndrome (del22q11.2) candidate gene and is required for pharyngeal and cardiovascular development. Tbx1-/- embryos have severe cardiac anomalies including a common arterial trunk.

DiGeorge syndrome patients have a high incidence of conotruncal defects including persistant truncus arteriosus and tetralogy of Fallot. We have shown that the common arterial trunk in Tbx1-/- embryos has an aorta-like phenotype associated with severe reduction of a subpopulation of second heart field progenitor cells normally contributing to myocardium at the base of pulmonary trunk. Underdevelopment of subpulmonary myocardium is thought to be the primary defect in human conotruncal defects like tetralogy of Fallot. Anomalous coronary artery patterning occurs in Tbx1-/- hearts. Semaphorin3c, encoding a neurovascular guidance molecule is expressed in a Tbx1-dependent domain in the subpulmonary myocardium. Disruption of the semaphorin signaling pathway during heart morphogenesis results in outflow tract defects and anomalies of the aortic arch arteries. Sema3c-/- embryos also display common arterial trunk with interruption of the aortic arch but coronary artery patterning appears normal.Here we present a comparative analysis of the evolution of common trunk in these two models and investigate potential genetic interaction between these genes.

Future subaortic and subpulmonary regions are prefigured in the E10.5 outflow tract. Using a candidate gene approach and microarray analysis at E10.5 we aim to identify additional genes expressed in subpulmonary myocardium that may contribute to conotruncal and coronary artery development.

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20 years of follow-up in 132 Senning procedures: late results.

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Objectives: Senning procedure was performed for the first time in France at Bordeaux University Hospital in 1965. Follow up have come out focusing on failing systemic right ventricle and rhythmic complication. The aim of this work is to identify prognosis factors of reoperation, arrhythmias and right ventricular dysfunction.

Methods: This single institution study is a retrospective analysis of 132 patients, with simple (105 patients) and complex (27 patients) transposition of the great arteries, after a Senning procedure between 1977 and 2004. The mean follow-up time was 19.5 ± 6.6 years. Conventional follow up and testing were performed in the same unit. An isotopic ventriculography or MRI was done for 70 patients to investigate the systemic right ventricular function.

Results: Operative mortality was 5.3 %. Late mortality was 9.6 %. 9 patients underwent a single reoperation and one needs heart transplantation. Actuarial survival rate was 91.5 %, 91 %, 89 % and 88 % at respectively 1, 5, 10, and 20 years. There is no statistically difference between simple and complex transposition for actuarial survival rate, maintaining permanent sinus rhythm or arrhythmias occurrence. After 20 years of 98 % of patients in simple transposition group have ejection fraction > 40 % versus 58 % in complex transposition ($p<0.001$). Risk factors of ventricular dysfunction were complex transposition ($p< 0.001$) and absence of cardioplegia ($p<0.001$). Last follow-up showed 91 % in NYHA class 1.

Conclusion: Imaging systemic right ventricular dysfunction (FEVD<40%) was yet uncommon at 20 years of follow-up but is not sufficient to predict a good response of this ventricle to stress and effort. Long term follow-up after the Senning operation shows frequent and increasing incidence of sinus node dysfunction and others arrhythmias: these complications are expected in double switch procedure.

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Long term results of chirurgical repair of aortic coarctation in Tunisia

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Introduction: Coarctation of the aorta (CoA) is a stenosis usually located in the isthmus of the descending aorta. Treatment consists of surgical or percutaneous removal of the obstruction and may present excellent immediate results. However, despite immediate good results, significant residual problems often persist. The aim of the study is to describe the presentation, treatment and long-term evolution of a population of 48 unselected consecutive patients with CoA in a single pediatric cardiology center.

Methods: This was a retrospective study of all patients with isolated CoA associated or not to either atrial or ventricular septal defects. RESULTS: The patients (n=48, 56,3% male) were diagnosed at a mean age of 84±109 months. The clinical presentation differed between patients aged less or more than two years, the former presenting with heart failure and the latter being asymptomatic with evidence of hypertension ($p < 0.01$). Treatment was surgical in all cases (32 end-to-end anastomosis). The mean age of patients was 94±109 months. There was two late deaths, in a mean follow-up of 8,6±7,7 years. Recoarctation occurred in 12 patients (25%). There are patients who currently have hypertension (17 at rest, 2 with effort), their mean age at diagnosis being older than the others (128 vs. 76 months; $p < 0.05$). Aortic aneurysms occurred in five patients (10,4 %). Aneurysm was associated to bicuspid aortic valve in 3 cases.

Conclusions: repaired CoA has a significant incidence of long-term complications, and should thus no longer be seen as a simple obstruction in the descending aorta, but rather as a complex pathology that requires careful follow-up after treatment.

January 14th, Friday 2011

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Are there indications for supraventricular re-entrant tachycardia in the youth?

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Radiofrequency (RF) ablation of paroxysmal supraventricular tachycardia (SVT) that could be an atrioventricular node re-entrant tachycardia (AVNRT) or AV reentrant tachycardia (AVRT) through a concealed accessory pathway (AP) is widely used in adults, but the indications remain controversed in children and teenagers. The purpose of the study was to evaluate the clinical and electrophysiological factors of indications of AVNRT and AVRT ablation in the youth and the results of the procedure.

Methods: 66 children or teenagers aged from 6 to 18 years (15±2) among 1099 patients consecutively recruited for SVT; they had a normal ECG in sinus rhythm; AVNRT or AVRT was confirmed at electrophysiological study (EPS). Patients with anterograde conduction through an AP were excluded.

Results: RF ablation was indicated in 26 children or teenagers (39 %) (group I) significantly less frequently than in adults (668/1033; 65 %) ($p < 0.0000$). 40 other children were not treated or received beta blockers (group II). Group I and II have the same age (15±2.6 vs 15±2.8). Male gender tended to be less frequent in group I (9/26; 35 %) than in group II (20/40; 50 %) (NS, 0.2). At EPS, group I had more frequently a concealed AP (13/26; 50 %) than group II (8/40; 20 %) ($p < 0.01$). The incidence of concealed AP in group I is higher than in adults (102 of 668 procedures;15 %) ($p < 000000$). In remaining children, ablation of slow pathway was performed in 13 children. There were no complications, but failures or reappearances of SVT after ablation were frequent (6/26; 23 %), more frequent in AVRT (4/13) than in AVNRT (2/13) (NS) and were related to the age (12.7±3 vs 16±2; $p < 0.002$). They tended to be more frequent than in adults (77/668; 11.5 %) ($p < 0.07$). In group II, 3 children continue the beta blockers and other children are not treated.

Conclusions: The indications of slow pathway remain rare in children. Most of the indications of SVT ablation in the youth which are rare, concern generally the children and teenagers with concealed AP. The results are almost as good as in adults, mainly for the slow pathway ablation and in teenagers.

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Is it still justifiable to treat right outflow track obstruction by bare metal stenting?

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Purpose: Right ventricular outflow track obstruction is an increasing concern in adult with congenital heart disease. Until recently, bare metal stenting (BMS) was the treatment of choice in this multi-operated population. Nethertheless pulmonary regurgitation resulting has not been studied in physiological condition yet. Furthermore, percutaneous pulmonary valve implantation (PPVI) is now possible. Our aim is to study changes in cardiac index occurring first after bare metal stenting and then after percutaneous valve implantation by magnetic resonance imaging performed under physiological condition.

Methods: seven consecutive patients (median age 17 years) with severe outflow tract obstruction underwent bare metal stenting followed a few weeks later by a percutaneous valve implantation. Magnetic resonance imaging (MRI) with left cardiac index based and net anterograde pulmonary blood flow was performed under physiological condition, before and after BMS and after PPVI.

Results: BMS significantly reduced outflow track obstruction (maximal velocity 3,19 versus 2,3 cm/s; $P<0,05$) with no significative change after PPVI (2,27 cm/s; P=1).After BMS, a free pulmonary regurgitation occurred (15 versus 38 %; $P<0,05$) and pulmonary cardiac index decreased (3,2 versus 2,4 l/min/ m$\sum$; $P<0,05$). After PPVI, there was no pulmonary regurgitation left (2 +/-1,4 %) and pulmonary blood flow showed a tendency to increase (2,4 versus 3,13 ml/min/ m$\sum$; P=0,07) while cardiac index significantly increased (2,7 versus 3,5 l/min/ m$\sum$; P=0,04) as well as left ventricular diastolic volume (70 versus 87ml; P=0,02).

Conclusion: Under physiological condition, bare metal stenting has a deleterious effect on cardiac index based on pulmonary blood flow and confirmed by left cardiac index. PPVI improves cardiac output eliminating the free pulmonary regurgitation. Impact of PPVI on right ventricle remodelling should be established with longer studies.

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Aorto-pulmonary anastomosis in tuberous sclerosis and cardiac tumor with severe right ventricular outflow tract obstruction (a case report)

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Primary cardiac tumors are rare with an estimated incidence of 0.27% in pediatric autopsies. The most common type of cardiac tumor identified in infancy and childhood is rhabdomyoma. We report a case of prenatal diagnosis of multiples cardiac rhabdomyoma at 29 weeks of gestation. Full-term delivery was straightforward without hydrops and dysrhythmia. In the immediate post-natal period, cyanosis appeared, and echocardiography showed multiple cardiac rhabdomyoma and severe right ventricular outflow tract obstruction (Image 1). Aprostadil perfusion was necessary. Aorto-pulmonary anastomisis was performed with success. The diagnosis of cardiac rhabdomyoma was confirmed histologically and tuberous sclerosis by molecular genetic analysis. Renal echocardiography was normal and cerebral MRI was not perfomed. Fourteen months later, neurodevelopment was normal. Echocardiography confirmed regression of the cardiac tumors with disappearance of the severe right ventricular outflow tract obstruction. ECG monitoring was normal.

Conclusion: The natural history of most cardiac rhabdomyoma is favorable with tumors regressing (completely or partially). Surgical resection of the tumors is required for severe ventricular outflow tract obstruction. Aorto-pulmonary anastomosis, as in our case with severe right ventricular outflow tract obstruction, is an alternative treatment because regression and even complete resolution of more than 80% of the tumors occurs during infancy and early childhood.

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Nonbacterial thrombotic (Libman-Sacks) endocarditis with mitral regurgitation in catastrophic antiphospholipid syndrome (a case study)

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